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Case Report

Primary Leiomyosarcoma of the Broad Ligament: A Case Report and Review of Literature

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Abstract

Primary leiomyosarcoma of the broad ligament is an extremely rare tumor with very few cases reported in literature. We herein report a case of postmenopausal lady, diagnosed pre-operatively on ultrasound and CT as a degenerated fibroid/ ovarian tumor. Post operatively histopathological diagnosis proved to be a leiomyosarcoma. The patient was treated with adjuvant radiotherapy with a dose schedule of 50 Gy in 25 fractions over 5 weeks. Patient is on monthly follow up since 6 months with no complaints.

Keywords: Leiomyosarcoma, tumor, postmenopausal, histopathological diagnosis, radiotherapy.

INTRODUCTION

Leiomyosarcoma (LMS) constitute approximately 5-10 % of all soft tissue sarcomas. By definition true broad ligament leiomyosarcomas occur on or in the broad ligament but are completely separated from and in no way connected with either the uterus or the ovary [1]. The histological criteria for the diagnosis of leiomyosarcoma are high mitotic index, cytological atypia and areas of necrosis. We hereby report a rare case of this highly malignant tumour in a postmenopausal female.

CASE REPORT

A 50-year-old post menopausal woman presented with dull aching pain and distension of lower abdomen since one week in the obstetrics and gynaecology department. On examination a non-tender mass 16 x13 cm in the left flank region and hypogastrium was felt with restricted mobility. On per vaginal examination uterus was not felt separate from the mass and uterine size could not be made out. Bilateral forniceal fullness was present. Abdominal and pelvic ultrasonography revealed a large complex cystic and solid lesion with vascularity in the pelvic cavity more on the left side. Uterus was atrophied in size and showed post menopausal changes. Ovaries were not visualized due to atrophy owing to postmenopausal status. CT scan of the abdomen and the pelvis (Figure 1-3) showed a 13.3 x 12.7 x 16.7 cm sized heterogeneously enhancing solid cystic lesion involving the pelvis, more on the left side with internal calcification. Lesion was abutting the uterus, sigmoid colon with compression of the left lower ureter causing, mild hydronephrosis and hydroureter. Fat planes were maintained between the mass, uterus and the rectum. Right ovary could not be determined separately. Serum carcinoembryonic antigen (CEA) and CA-125 were mildly elevated. With these background features on ultrasound and computed tomography scan, possible diagnosis of either a degenerating broad ligament fibroid or an ovarian tumor was made. The patient was referred to the Gynaec- Oncology OPD where total abdominal hysterectomy with bilateral salpingo-oophorectomy with excision of left broad ligament mass was done. Left ruptured para-tubal broad ligament mass measuring 10 x 7.5 x2.0 cms in size was removed. Gross examination of the specimen of the left broad ligament mass showed a well encapsulated tumour with solid hemorrhagic areas and few cystic areas with a smooth cyst wall. Microscopy [Fig. 4, 5] showed high grade spindle cell sarcoma with epithelioid differentiation. There was marked pleomorphism and necrosis. Mitotic figures seen were 10/10 HPF. Immune-histochemistry (IHC) showed vimentin, desmin, actin and EMA positive and S-100, AE1, PAX 8, PLAP, CK 7, HMB 45 negative. The IHC diagnosis of high grade leiomyosarcoma with epithelioid differentiation was made.

Since very few cases have been reported, the staging and management of leiomyosarcoma of the broad ligament are still based on criteria used for uterine leiomyosarcoma. Prior studies focusing on LMS have reported several clinical and pathological

characteristics to be prognostic for clinical outcomes. Specifically, advanced age (>55 years), high tumor grade, larger tumor size (>5 cm), high mitotic index (≥ 15 per high power field), and omission of bilateral oophorectomies have been correlated with worse outcomes.

The patient presented a month later in the radiation oncology OPD for adjuvant treatment. The patient was prescribed adjuvant pelvic radiotherapy with a dose schedule of 50 Gy in 25 fractions over 5 weeks at 200cGy per fraction in two parallel opposed fields (antero-posterior/ postero-anterior) of size 16cm x 17 cm with each field receiving 100cGy.

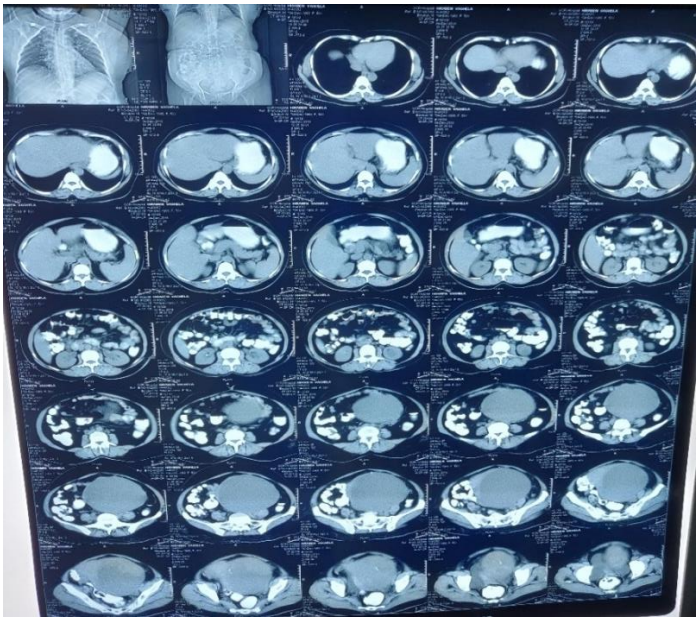

Fig-1:

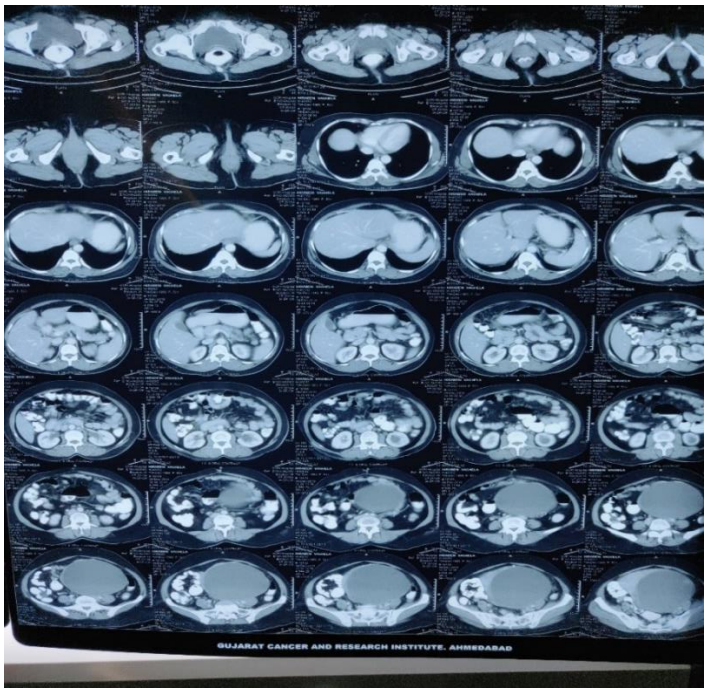

Fig-2:

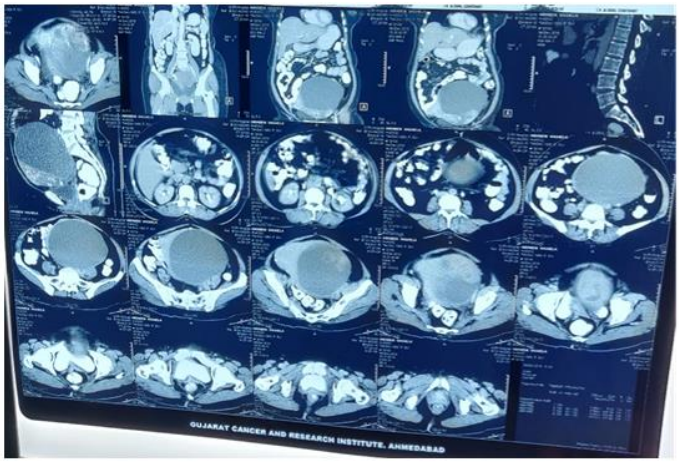

Fig-3:

Figure 1 to 3 CECT abdomen and pelvis (axial view / sagittal view) shows a well defined heterogeneously enhancing lesion measuring 13.3 x 12.7 cm with central necrotic area in left adnexa. Fat planes with uterus are distinct

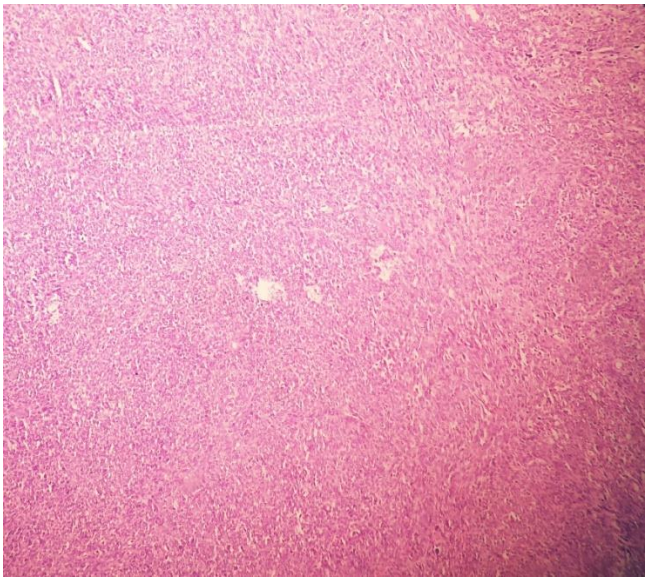

Fig-4: H&E (10 ×) section

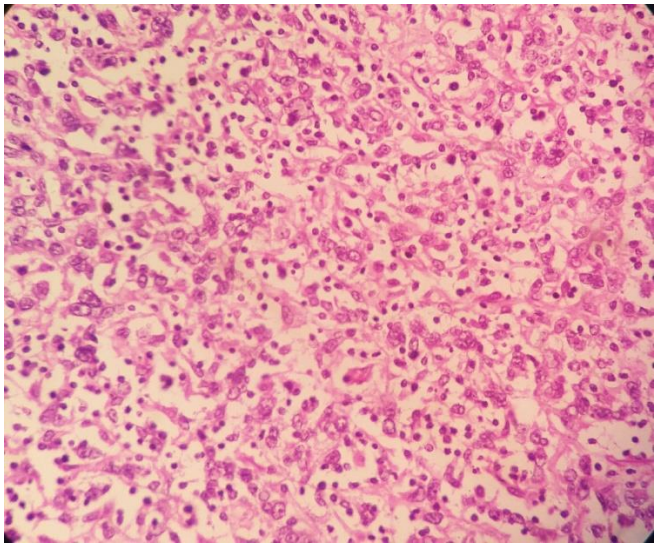

Fig-5: H&E (40 ×) section shows intersecting fascicles of malignant spindle cells with adjoining area of necrosis

DISCUSSION

Till now only 24 cases of primary leiomyosarcoma of the broad ligament have been reported in the literature published in English [21]. It is a very rare tumour and a rapidly progressive one. It is also a highly malignant gynaecological tumour [4]. It is a malignant mesenchymal tumour of cells showing smooth muscle differentiation. Distinguishing between leiomyoma and leiomyosarcoma is difficult because of the similar presenting features since leiomyoma can undergo secondary changes which include degeneration, necrosis, haemorrhage, and rarely sarcomatous change. These tumours commonly occur in postmenopausal women. The reported cases of primary malignant tumours of the broad ligament are leiomyosarcoma, endometrial stromal sarcoma, malignant fibrous histiocytoma and carcinosarcoma [20]. The clinical manifestations of these cases are non-specific symptoms and signs that include abdominal pain, distension, nausea, constipation, and malaise. Broad ligament fibroids are associated with pseudo Meigs' syndrome and they can produce elevated serum marker CA-125 levels, leading to diagnostic confusion with metastatic ovarian carcinoma [22]. Pelvic imaging techniques cannot reliably differentiate between a benign leiomyoma and a sarcomatous degeneration as both can undergo central necrosis. USG features of mixed echogenic parts, central necrosis, and colour Doppler findings of irregular vessel distribution, low impedance to flow, can also be seen in a benign leiomyoma. As in our case the USG findings mimicked fibroid with degenerative changes or an ovarian tumor. Due to the anatomical close proximity, these broad ligament fibroids may be confused with adnexal neoplasms.

The earliest criteria laid down was by Gardner *et al*., [1] in 1977 which established that the disease should be ''completely separated from and in no way connected with either the uterus and the ovary.'' The other systems used for grading of soft tissue sarcomas are the NCI (National Cancer Institute) and FNCLCC
(Fédération Nationale des Centres de Lutte Contre le Ca ncers) systems. The NCI system uses a combination of histological type, cellularity, pleomorphism and mitotic rate for attributing grade 1 or 3. The FNCLCC grading system is based on a score obtained by evaluating three parameters: tumour differentiation, mitotic rate (0–9, 10–19 & > 20 mitoses/10 HPFs) and the amount of tumour necrosis (< 50% tumour necrosis and > 50% tumour necrosis). According to both these systems, leiomyosarcoma is classified as low, intermediate and high grade [20]. Zaloudek and Norris [2] consider separately mitotic figures (equal to or more than five versus less), cellularity of the tumour (hypercellular versus normocellular), and nuclear atypia (yes versus not) as the most important criteria to differentiate leiomyosarcoma from leiomyoma. Coindre et al. [15] assume that mitotic figures, tumor differentiation, and tumor necrosis should be considered together. The Stanford study [3] was the first to appreciate that necrosis in a uterine smooth muscle tumour was of crucial importance. In the absence of cell necrosis, the diagnosis of leiomyosarcoma needs diffuse moderate to severe cellular atypia and more than 10 mitoses/10 HPFs [13]. The criteria described by Bell *et al*., [12] and refined by Hendrickson and Kempson [3] for diagnosis of uterine smooth muscle neoplasms are: Degree of cytological atypia (none to mild or moderate to marked), Presence or absence of coagulative tumour cell necrosis, Mitotic index if moderate/severe atypia is present without necrosis

The differential diagnosis includes masses from ovarian origin, either benign or malignant, broad ligament cyst, lymphadenopathy and tubo-ovarian masses. CT scan does not reliably diagnose between leiomyomas and leiomyosarcomas. Lee *et al*., [23] proposed "ovarian vascular pedicle" as a way of differentiating ovarian neoplasms from adnexal lesions by single detector helical CT scan. Leiomyosarcoma and leiomyoma can be differentiated microscopically by the following criteria. Presence of 5 or more mitotic figures per 10 high power field, nuclear atypia, hyper-cellularity is required to diagnose a leiomyosarcoma. The prognosis for women with uterine sarcoma primarily depends on the extent of the disease at the time of diagnosis and mitotic index [24].

There is wide variation in the management practices of this uncommon tumour but initial treatment is same as that of uterine leiomyosarcoma, i.e., total abdominal hysterectomy and bilateral salpingo-oophorectomy. Pelvic lymph node dissection is debatable. The imaging and microscopic pattern play a crucial role in defining overall prognosis and need for adjuvant therapy. Pelvic irradiation therapy has been used for adjuvant treatment of uterine leiomyosarcomas as radiation therapy and has been shown to decrease the pelvic locoregional relapse rate.

Since very few cases have been reported, treatment of broad ligament leiomyosarcoma is based on the criteria used for uterine leiomyosarcoma [12]. Standard management for uterine sarcoma includes total abdominal hysterectomy and bilateral salpingo-oophorectomy [14], but pelvic and para-aortic lymphadenectomy is recommended only for carcinosarcoma and not for leiomyosarcoma, despite the fact that some cases of the latter show a higher incidence of nodal involvement. Occasionally, the tumor is large, adheres to surrounding structures, and has areas of haemorrhage or necrosis. The role of frozen section in the diagnosis can be omitted, but in our case, frozen section revealed an accurate diagnosis. In nulliparous, pre-menopausal women or in the case of a tumor in early its stages, the ovaries should be

conserved. Adjuvant chemotherapy with or without radiotherapy can be used in selected cases. Some authors believe that when mitoses are less than 10 per 10 HPF, no further therapy is needed after surgical intervention [10]. There is little evidence to support use of adjuvant chemotherapy for any gynaecological sarcoma except carcinosarcomas, although doxorubicin/ epidoxorubicin with ifosfamide has been used for leiomyosarcomas, in conjunction with pelvic radiation after chemotherapy in selected cases. Most of the tumours with high mitotic figures reported in literature have been managed with adjuvant postoperative combination chemotherapy and/or radiation. Recently, fixed-dose gemcitabine (900 mg/m^2 on day 1 and 8) used along with docetaxel (100 mg/m2 on day 8) has been shown to achieve high objective response rates as a first line therapy in metastatic uterine leiomyosarcomas [16].

CONCLUSION

Broad ligament leiomyosarcoma is difficult to diagnose with clinical and radiological studies, histopathology plays a prime role for making the diagnosis. Because of their rarity, they are not suitable for screening. Surgery is the treatment of choice with postoperative radio and chemotherapy in recommended cases. In view of the unpredictable behaviour of this rare tumor and the empirical criteria used for its management, complete surgery supplemented with adjuvant chemotherapy or radiation and a close follow up for recurrence would help optimize the chances of disease-free survival.

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